

Book Review: Rongioletti F, Smoller BR (eds.). Clinical and Pathological Aspects of Skin Diseases in Endocrine, Metabolic, Nutritional and Deposition Disease. New York: Springer; 2010.

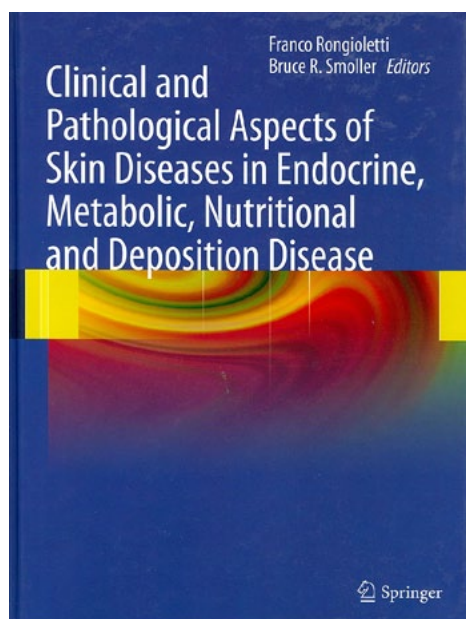
Reviews by Christine Ko, M.D. and François Milette, M.D. with response from the authors
Editorial comment by Mark A. Hurt, M.D., Book Review Editor

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Rongioletti F, Smoller BR (eds.). *Clinical and Pathological Aspects of Skin Diseases in Endocrine, Metabolic, Nutritional and Deposition Disease*. New York: Springer; 2010. 195 pp with index. ISBN 978-1-60761-180-6.



Review by Christine Ko, M.D.

This is an ambitious undertaking, as suggested by the lengthy title, and the authors successfully cover endocrine, metabolic, nutritional, and deposition disorders. The book is all the more thorough as there is seamless assimilation of clinical, histologic, and radiologic information for these diseases. The

aims are fulfilled with 1) concise text, 2) atlas-like inclusion of over 174 clinical photos and 116 histopathologic photomicrographs, 3) insight into the clinical appearance and course of disease, 4) detailed histopathologic descriptions, 5) therapeutic options, and 6) bulleted key points. This book should appeal to all dermatopathologists, as it is compact, richly illustrated, and easy to read.

Some minor criticisms include a need for white balancing many of the histopathologic photomicrographs and for improvement of brightness and color of the clinical photographs. Despite this, the figures more than adequately depict the numerous diseases summarized in this book. In particular, some of the images not to be missed include beautiful clinical examples of necrobiosis lipoidica (Fig. 2.1), pretibial myxedema (plaque-type) (Fig. 3.2), acromegaly (Fig. 4.1), eruptive xanthoma (Fig. 6.4), fixed drug (Fig. 14.6), and primary systemic amyloidosis (Fig. 17.4). This atlas further delves into variants of granuloma annulare (chapter 2), pretibial myxedema (chapter 3), xanthomas (chapter 6), and mucinoses (chapter 18). While this is a strength, it also is a minor weakness, as the appetite is whetted to see even more images of rare variants like discrete papular lichen myxedematosus and pure nodular lichen myxedematosus.

The authors are recognized experts in the subjects covered in this book, and even while achieving their broad aims, the authors include valuable pearls throughout the text.

Some involve differentiating certain disorders: for example, acne secondary to hypercorticism lacks comedones and cysts (unlike acne vulgaris), siderosis around eccrine glands may be specific for idiopathic hemochromatosis, and the linear array of papules is a clue for scleromyxedema as opposed to scleredema or scleroderma. A treatment pearl is using the hyperpigmentation of Addison disease to monitor disease activity. As the authors' expertise shines through with comprehensive coverage of the myriad diseases, it would be of value to know their opinion on issues like:

A) Lichen myxedematosus – how frequently is it associated with paraproteinemia or other systemic findings [1]? Should all cases be screened and, if so, just once?

B) Reticular erythematous mucinosis – the literature suggests many cases are associated with autoimmune disorders [2]. Is the author's experience different?

C) Calciphylaxis – how often is it **not** in the setting of renal failure [3, 4]? Is there a better prognosis if **not** truncal?

Overall, this book succinctly achieves a wide-ranging look at endocrine, metabolic, nutritional, and deposition disease. Concise text accompanies numerous photographs and photomicrographs, enabling the reader to get a handle on these diseases clinically and histopathologically. This is a worthy read for anyone interested in these disorders.

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Review by François Milette, MD

This book is presented to its reader as “a concise atlas and text for the practicing dermatologist, pathologist, dermatopathologist, endocrinologist, and internist.” In this context, what should we expect from it?

Certainly an *atlas* should be complete and well-illustrated, whereas a *text* (or textbook) should be clearly and logically written. If it meets those requirements, it has a chance of being practically useful. Last – and this is related to completeness of covering of the subject – to be worth its price, it should add to what is available in other textbooks already published.

Therefore, I have retained three criteria to evaluate in my review of this book: completeness, image quality, and logic and clarity of the approach presented.

Concerning completeness, suffice it to say that an atlas pertaining to, among other things, metabolic diseases with cutaneous manifestations should include lysosomal diseases (mucopolysaccharidoses, glycogenoses, sphingolipidoses, cystinosis, etc.) that do not appear in this book but can be found in other textbooks [1], therefore more complete at least on this point. Those diseases are extremely rare, of course, but this is no excuse to neglect them, as lipoid proteinosis, which is discussed at length in a chapter devoted to it alone in this book, can hardly be considered frequent.

As for the quality of the images, great efforts have been made to include numerous clinical photographs and histological microphotographs. The quality of these images, however, is very unequal. Some are crystal clear but, unfortunately, others are suboptimal and look as if they had been taken through filters of various colors: blue, green, yellow, pink, etc. With the technology available today, it should have been relatively easy to standardize the quality of the images. One wonders, by the way, why the photograph on page 185 occupies the whole page and is the only photograph in the whole book to do so.

Concerning the logic of the approach, things start badly, as no introduction is offered to define the various categories of diseases discussed in the book. One may argue that everybody knows what endocrine, metabolic, nutritional, and deposition diseases are, but the authors themselves admit rightly that the categories as they use it are not exclusive, therefore questioning the pertinence of their enterprise. Moreover, the warning placed in the preface that some diseases will appear in more than one category made this reader uneasy and suggests that some confusion exists in the organization of this book. For me, this was confirmed by the reading of it, which I found very hard to complete. The authors suggest that that hormones and nutrition influence metabolism profoundly and any “deposition” is the result of faulty metabolism, the incapacity to catabolize the material

deposited. Therefore, the inclusion of the entities discussed in one or another category can only be arbitrary, and it is.

One of the consequences that results from classifying into undefined and overlapping categories is that some diseases are classified in a manner that appears simply nonsensical to a naïve reader. If hyperlipidemia is easily understood as a metabolic disease resulting from inherited or toxic faulty metabolism of lipids, it is hard to understand how obesity can be included in the same group of diseases and not considered nutritional! Isn't obesity the stereotypical example of nutritional disease, or does it result, as in the mind of the authors, from a faulty metabolism?

And what about glucagonoma syndrome (GS) and necrolytic migratory erythema (NME)? Glucagonoma is a neoplasm and, therefore, logically, GS and NME, the physiopathology of which is unknown, are classical paraneoplastic syndromes as the authors themselves admit in the very first sentence of their chapter 16. So why do they discuss it as a nutritional disease? And why isn't glucagonoma discussed in the chapter on endocrine or pancreatic diseases or, if need be, as a "metabolic" disease?

This book raises more problems than it offers solutions, and many, many other questions could be enumerated here, but this is only a book review and no place for such lengthy development.

In this work, the authors place the Latin phrase, *repetita iuvant*, repetition is useful. By this, they want to justify the presence of some entities in more than one chapter as intended to offer "different perspectives." It is not, however, repetition to which the reader is exposed; it is to redundancy and to arbitrary, incomplete and fairly illogical classification of rare diseases. After going through this book, my comprehension of the subjects addressed by it was not made clearer, and in response to the use of the Latin phrase *repetita iuvant*, I would say that if it is true that repeating the same phone number three times helps in remembering it, three different numbers told in a row are certainly not easier to learn.

Lastly, the very title of this book sounds bizarre: *Clinical and pathological aspects of skin diseases [sic] in endocrine, metabolic, nutritional and deposition disease [sic]*. In short: *diseases in disease*. One immediately feels uneasy. If some entities described, for instance, necrobiosis lipoidica, are indeed diseases of their own that may occur alone or in association with another disease (in this case diabetes), others can simply not be described as diseases. Mucinosis and calcinosis, for example, are not diseases but histopathologic patterns occurring in many different contexts, e.g., neoplastic, endocrine, inflammatory, etc. It is no surprise; therefore, that confusion arises from the reading of this book confounding diseases and patterns. A more adequate title could have read *Cutaneous manifestations of endocrine, metabolic, nutritional and deposition diseases: Clinical and pathological aspects* and

perhaps would have helped the work to be more coherent.

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Drs. Rongioletti & Smoller respond to Dr. Milette's review

Our main goal was not to write a complete and exhaustive compendium. Rather, it was to write just a concise atlas-text to transfer to the practicing dermatologist, pathologist, endocrinologist, and internist all of our experience and our photographic and graphic materials on some topics that are often covered scantily, receive little emphasis, or are found scattered in different sections of the general textbooks of dermatology or dermatopathology.

This is a first edition where, of course, there is ample room for improvement and all criticisms are welcome and really appreciated. Milette's criticisms are best divided into constructive and irrelevant.

Constructive criticism

The criticism for not including lysosomal disorders is valid. However, these diseases belong largely to the pediatric sphere that is not a strict part of our expertise. We understand that all this does not necessarily follow from the title of the volume, and actually may leave some readers unsatisfied. In fact, we have discussed adding "in adults" to the title (which is already perceived as long). If a next edition is taken into consideration, it will be our first concern to add a chapter devoted to lysosomal storage diseases.

We agree that some pictures are not of outstanding quality. It is not always easy to get good clinical and histological photographs from uncommon diseases. Some pictures were old, difficult to reproduce, and supplied from the archives of the departments; the individuals who wrote the chapters supplied some others. Of course, we know that is Editor's task to ensure that the image quality is optimal. We will take heed of this suggestion for the future.

Irrelevant criticisms for improvement

Classification: There is no perfect classification on which all can agree. Classification, however, is a fundamental process and a necessary starting point in the study of the natural history of disease. We think that there is nothing wrong to

classify obesity as a metabolic disease, and we are not alone in this position [1]. Moreover, *isn't it true* that the association of obesity with hypertension, glucose intolerance, and lipid metabolism disorder is diagnosed as metabolic syndrome [2]? The same holds for diabetes, which is classified both as a metabolic or a nutritional disease in various textbooks and articles. Necrolytic migratory erythema has been included in the chapter of “nutritional” for two reasons: it is considered pathogenetically due to a relative deficiency of zinc and essential fatty acids because the tumor reduces the amount of albumin that normally carries it around the body, and its histological features are similar to acrodermatitis enteropathica and pellagra, which are clearly nutritional disorders. Necrolytic migratory erythema has also been seen in the absence of a glucagon-producing tumor and may be associated with various systemic diseases, such as celiac disease, ulcerative colitis, etc. Thus, its inclusion in the chapter of pancreatic disease or paraneoplastic disorders could also have been questioned.

Concerning the repetition of just a few disease in different chapters, this is not a matter of redundancy or arbitrary and incomplete classification of rare diseases, but, on the contrary, it reflects how metabolic, endocrinologic, nutritional, and deposition diseases may have overlapping features presenting with the same skin manifestation. For instance, pretibial myxedema can be considered both as a skin manifestation of thyroid disease and also classified among the primary cutaneous mucinoses; the same is true for macular or lichen amyloidosis that can be considered both as a skin manifestation of MEN 2A and also classified among the primary cutaneous amyloidoses. We think that there is nothing obscure or confusing to discuss a disease that is always the same but occurs in different backgrounds and eventually from distinct perspectives. Of course, the more the reader is familiar with this field, the higher and clearer is the perception and the understanding of the different diseases. This book, in fact, is not for absolute beginner.

The criticism of the words “skin diseases” in the title and the suggestion that they should be replaced by “cutaneous manifestations” is an example of an irrelevant criticism without any practical consequence. “Disease” as paraphrased according to the Merriam-Webster dictionary is a condition of the living animal or of one of its parts that impairs normal functioning and is typically manifested by distinguishing signs and symptoms. Thus, we don't understand why the term cannot be applied in our setting. Many of the illustrated skin conditions are “disease” per se and not only skin manifestations of systemic conditions. Incidentally, we consider cutaneous mucinoses and cutaneous calcifying disorders not as simple histological patterns but as a group of different deposition disorders that are heterogeneous. An accurate knowledge of their clinicopathologic features is essential in

order not to miss some important underlying association (scleromyxedema and monoclonal gammopathy, calciphylaxis in renal disease, etc).

In conclusion, we believe constructive criticism is welcome and worth voicing and may contribute to the improvement of our book. Spurious criticism goes nowhere and sometimes may mask an underlying lack of robust arguments.

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Final comment by Mark A. Hurt, MD

I thank Drs. Ko and Milette for their reviews of this book, and I thank also Drs. Rongioletti and Smoller for considering and responding to Dr. Milette's review.

As Drs. Rongioletti and Smoller are both editors and authors, they are not the *only* authors of the book. There is a total of 23 authors, including the editors, and they are positioned principally in Europe and the United States, the exception being two authors from Mexico. Of the 23 chapters in the book, all but nine are written by authors other than the editors.

In their *Preface*, Drs. Smoller and Rongioletti lay out the reason and purpose of their book: “Dr. Rongioletti has been studying, diagnosing, treating, and researching mucinoses and deposition disorders for years and this work represents his dream of consolidating his amassed experiences into a concise atlas and text for the practicing dermatologist, pathologist, dermatopathologist, endocrinologist, and internist.” Furthermore “Dr. Smoller has also been actively interested in cutaneous pathology as a window for systemic diseases and his written extensively on this topic.” Finally

“It is the hope of the authors that each chapter will provide insights into the clinical appearance and course of the diseases presented, followed by a review of the pathogenesis for each disease, and a close inspection of the histopathologic changes and any accompanying special studies that might be required to establish a diagnosis.”

This is a book that is of standard journal size, is relatively thin considering the subject matter, at 194 pages including the index, and the paper is of reasonably good quality. The editors divide the book into four major parts, as follows:

- I. Cutaneous Endocrine Disease (classified by the diseased organ)
- II. Cutaneous Metabolic Disease (classified by the product of the abnormal metabolism)
- III. Cutaneous Nutritional Disease (classified by the product of deficiency or excess without respect, necessarily, to cause)
- IV. Cutaneous Deposition Disease (classified by the product of deposition)

The authors present the chapters within each part as if they were separate small reviews as though in a journal. Each begins with a section of “key points” similar to an abstract, except organized by bullet points, which I find to be a useful feature. The organization of subject matter within each chapter is by a number outline, adding additional sub-points as needed; there is no strict separation into labeled sections of “clinical presentation,” “histopathological findings,” etc., even though those descriptions are found in the paragraphs of the text. Furthermore, the authors augment the text with ample photographs of somewhat uneven quality that refer back to the text thus allowing one to find where the authors discuss the histopathological findings. The text flows easily from well-constructed sentences within concise paragraphs. Although most of the abbreviations are relatively easy to grasp from the text, a key to abbreviations in the front of each chapter might be a consideration for any future editions.

Although one can criticize the authors for some of the photographs in this book, I am amazed at how many very good to excellent photographs *are* included, especially given the rarity of some of the conditions. Minor improvements of standardization of photographic hue and contrast will serve to make the second edition better.

I offer one consideration for any future edition of this book, and it is a difficult addition: a diagnostic algorithm linked to the text. Although this is a difficult task, and it perhaps is unfair to ask so much of the authors, it is enormously beneficial to have an algorithm when confronted with specific, concrete histopathologic findings in the daily work of developing a differential diagnosis for the purpose of rendering a specific diagnosis. Thus, a three-pronged approach – an algorithm, a text (with atlas), and a well-developed index –

should serve well the second edition of this book. Until then, I will become a serious student of *this* version.

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